

Severe tricuspid stenosis presenting as tricuspid atresia

Echocardiographic diagnosis and surgical management

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SUMMARY Two cases of pulmonary atresia with intact ventricular septum and severe tricuspid stenosis are described in which the initial angiographic diagnosis was tricuspid atresia and pulmonary atresia. Two dimensional echocardiography showed the features of an imperforate tricuspid valve because in each case contrast echocardiography failed to show anterograde flow from the right atrium to right ventricle. Successful radical surgical repair was achieved in one patient by performing a tricuspid valvotomy and inserting an external valved conduit between the hypoplastic right ventricle and the main pulmonary artery. In the second case, an infant died four weeks after tricuspid valvotomy and right ventricular outflow tract reconstruction.

The majority of hearts in which selective right atrial angiography shows no direct communication with a ventricular chamber are considered to be examples of tricuspid atresia. Most cases have absence of the right atrioventricular connection, the cardinal anatomical feature being a lack of any potential communication between the floor of the right atrium and the ventricular mass. A similar clinical picture arises when there is an imperforate right atrioventricular valve or severe stenosis, but the anatomical distinction between the three is not readily made by angiography. The description of the radical repair of tricuspid atresia by Fontan and Baudet¹ has made accurate analysis of the atrioventricular junction an essential prerequisite to surgery.

Case reports

CASE 1

The patient was a full term infant who had central cyanosis on the first day of life. At the age of 2 years she was severely cyanosed. There was a normal first heart sound, a single second sound, and a grade 2 short ejection systolic murmur heard maximally in the second and third intercostal spaces at the left sternal edge. The liver was palpable on the right. The electrocardiogram showed sinus rhythm with a mean frontal QRS axis of +150, biatrial enlargement with a dominant R wave in V1 of 25 mm, and an RS ratio of one in lead V6 with an R wave of 22 mm. The chest radiograph showed mild cardiomegaly with oligemic lung

fields, a left sided aortic arch, and a left sided stomach. Right heart catheterisation was selective right atrial angiography, and left ventriculography was thought to show tricuspid atresia and pulmonary atresia, though no anterior rudimentary right ventricle was visualised. A right Blalock-Taussig anastomosis was performed with good effect.

At the age of 5 years repeat cardiac catheterisation was performed because the patient was developing increasing cyanosis and exercise intolerance. Good sized main, left, and right pulmonary arteries filled via the right Blalock-Taussig shunt. Additional investigations by two dimensional echocardiography showed the features of an imperforate right atrioventricular valve which was supported by tensor apparatus and was seen to balloon into a small anterior right sided ventricular chamber during diastole using a peripheral venous injection of 5% dextrose. Contrast echocardiography failed to show any anterograde flow from the right atrium to the right ventricle (Fig.). The heart was now considered to be an example of atrioventricular concordance, with a perforate left and imperforate right atrioventricular valve, and pulmonary atresia.

At operation the tricuspid valve leaflets were found to be almost completely fused apart from a pin-hole orifice. The subvalvar apparatus was present, if somewhat rudimentary, and was considered to be sufficiently well developed to produce a competent valve after valvotomy. An incision was made in the infundibulum of the right ventricle. Pulmonary valve atresia with an intact ventricular septum was confirmed. A tricuspid valvotomy was therefore

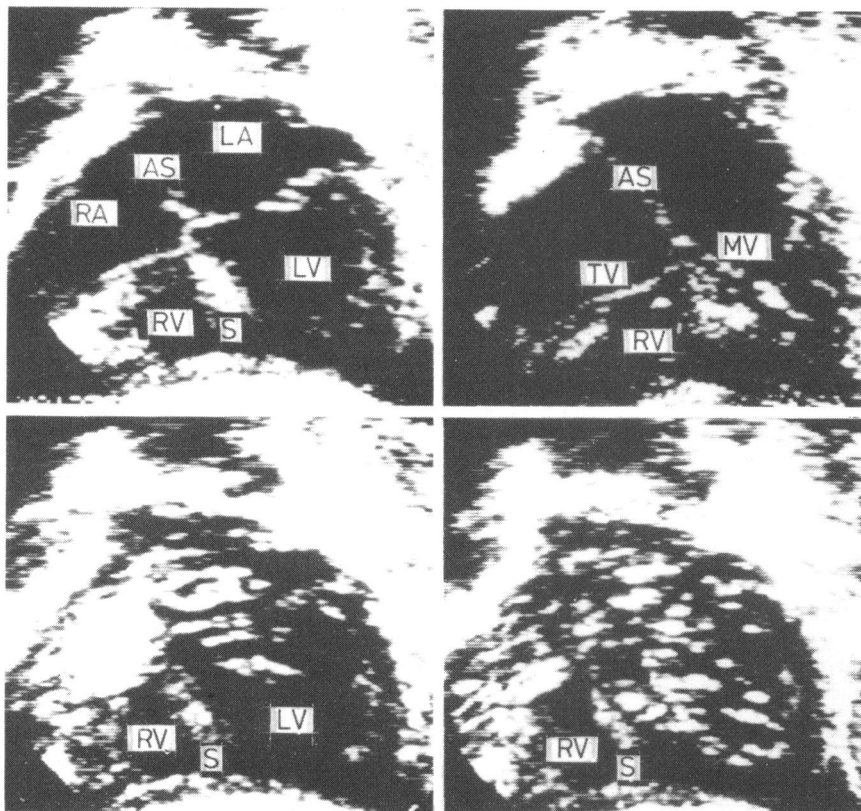


Fig. Serial contrast echocardiograms from a patient with atrioventricular concordance and an imperforate tricuspid valve (TV). The upper right and upper left stop frames show systole and diastole, respectively, and show that the tricuspid valve fails to open. In the lower right frame contrast echoes are seen in the right atrium (RA) crossing the atrial septum (AS) to the left atrium (LA). The lower left frame shows that the right ventricle (RV) fails to be opacified by contrast echoes. (S, ventricular septum; LV, left ventricle; MV, mitral valve).

performed and an external conduit was placed between the right ventricle and the pulmonary artery. The early postoperative period was complicated by right heart failure and tricuspid regurgitation, and diuretic treatment was required for six months. At present the patient has no symptoms, a normal exercise tolerance, and no clinical evidence of tricuspid valve regurgitation. Two dimensional echocardiography shows a tricuspid valve that opens well and there has been an increase in size of the right ventricle. The chest radiograph shows a cardiothoracic ratio of 0.48 and normal pulmonary vascular markings.

CASE 2

A male infant was found to be cyanosed on the first day of life. Physical examination showed central cyanosis and tachypnoea. The peripheral pulses and cardiac impulses were normal. There was a normal first heart sound and a grade 3 continuous murmur audible at the second left intercostal space.

On the chest radiograph the heart was slightly enlarged, the pulmonary vascular markings were decreased, and the aortic arch and stomach were left sided. The standard electrocardiogram showed sinus rhythm, a mean frontal QRS axis of -60° , right atrial enlargement, and left ventricular dominance. The two dimensional echocardiogram showed atrioventricular concordance, with an imperforate tricuspid valve and moderately hypoplastic right ventricle. Contrast echocardiography, using a peripheral venous injection of 2 ml 5% dextrose, failed to show any anterograde flow from right atrium to right ventricle. At cardiac catheterisation selective right atrial and left ventricular angiography demonstrated the features of tricuspid atresia but failed to outline the right ventricle. There was pulmonary valve atresia but the pulmonary arteries were of normal size, pulmonary blood flow being maintained by a large persistent ductus arteriosus.

The diagnosis was considered to be atrial situs solitus, atrioventricular concordance, ventriculo-

arterial concordance, imperforate tricuspid valve, hypoplastic right ventricle, and pulmonary valve atresia. At operation the tricuspid valve was found to have a small central orifice. An open tricuspid valvotomy was performed and the tensor apparatus of the valve was identified. The pulmonary artery was opened in its long axis and confirmed that the pulmonary valve was imperforate. An excision was extended from the pulmonary artery to the body of the infundibulum and the outflow tract was enlarged with a patch of Impre. The postoperative period was complicated by severe right heart failure. Two dimensional echocardiography showed poor mobility of the tricuspid valve leaflets. The infant developed jaundice and a severe chest infection and died four weeks after operation.

Discussion

In the majority of patients presenting with the clinical features of classical tricuspid atresia there is complete absence of the right atrioventricular connection and no potential communication between the right atrium and a ventricular chamber, sulcus tissue interposing between them.² Nevertheless, cases have been described in which the right atrium is in potential communication with an anterior rudimentary right ventricle through an imperforate tricuspid valve.³ It seems from our experience that the distinction between an imperforate and severely stenosed tricuspid valve cannot readily be made by selective right atrial angiography or contrast echocardiography because each technique may fail to show antegrade flow from the right atrium to the right ventricle. Imperforate or severely stenosed valves may also exist in univentricular hearts in which the type of atrioventricular connection is through two valves and the mode of connection is via a functionally imperforate right and perforate left atrioventricular valve.⁴

Imperforate right atrioventricular valves have been described briefly by several authors and there have been cases reported with coexisting Ebstein's anomaly.⁵ We have recently described the two dimensional echocardiographic features of imperforate valves in double inlet univentricular hearts, emphasising that the distinction from absence of the corresponding atrioventricular connection can readily be made.⁴ The cornerstone of the diagnosis of an imperforate valve is the identification of a thin mobile membrane between an atrium and a ventricle, and the recognition of the tensor apparatus of the valve. In the majority of cases the tensor apparatus is hypoplastic and therefore its delineation by echocardiography is not always possible.

We have described two cases of atrioventricular concordance with a functionally imperforate tricuspid valve possessing a pin-hole orifice, treated by tricuspid valvotomy. The distinction of this anomaly from absence of the right atrioventricular connection was dependent upon an accurate assessment of the atrioventricular junction by two dimensional echocardiography. From a surgical standpoint, some hearts with an imperforate right atrioventricular valve would be unsuitable for tricuspid valvotomy. These include double inlet univentricular hearts and cases with atrioventricular concordance in which the tensor apparatus or the right ventricle are hypoplastic. While the majority of patients presenting with the clinical features of tricuspid atresia will have absence of the right atrioventricular connection, a small number will have an imperforate valve. From our own experience it is evident that some of the latter may be managed by tricuspid valvotomy rather than by the Fontan procedure. In our second case, palliative treatment by a shunt procedure may have been the preferable initial treatment, tricuspid valvotomy and right ventricular outflow tract reconstruction being reserved for later. Except in univentricular hearts, imperforate tricuspid valves occur so rarely that whenever this diagnosis is made echocardiographically the alternative possibility of severe tricuspid stenosis should be strongly suspected, for under these circumstances valvotomy may be feasible.

References

- 1 Fontan F, Baudet E. Surgical repair of tricuspid atresia. *Thorax* 1971; **26**: 240-8.
- 2 Anderson RH, Wilkinson JL, Gerlis LM, Smith A, Becker AE. Atresia of the right atrioventricular orifice. *Br Heart J* 1977; **39**: 414-28.
- 3 Dickinson DF, Wilkinson JL, Smith A, Anderson RH. Atresia of the right atrioventricular orifice with atrioventricular concordance. *Br Heart J* 1979; **42**: 9-14.
- 4 Rigby ML, Anderson RH, Gibson D, Jones ODH, Joseph MC, Shinebourne EA. Two dimensional echocardiographic categorisation of the univentricular heart. Ventricular morphology, type, and mode of atrioventricular connection. *Br Heart J* 1981; **46**: 603-12.
- 5 Rao PS, Jue KL, Isabel-Jones J, Ruttenberg HD. Ebstein's malformation of the tricuspid valve with atresia. *Am J Cardiol* 1973; **32**: 1004-9.

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